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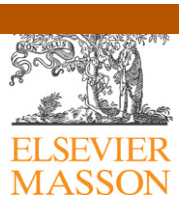

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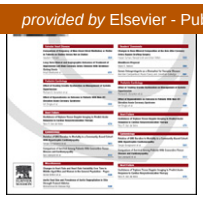

IMAGE

An unusual case of hypertensive cardiomyopathy

Un cas inhabituel de cardiomyopathie hypertensive

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MOTS CLÉS
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A 32-year-old man, with a medical history notable only for severe hypertension, was referred to our hospital for hydrocephaly, secondary to a voluminous, hypervascularized, right tympanic tumour encasing the jugular vein and extending to the posterior fossa (Fig. 1A).

On admission, the patient was dyspnoeic. Blood pressure was 180/93 mmHg and heart rate was 117 beats/min. Plasma brain natriuretic protein concentration was 2200 pg/mL. Echocardiography showed left ventricular (LV) dilatation (end-diastolic diameter 70 mm) and non-obstructive global LV hypertrophy (LV mass index 200 g/m^2). LV ejection fraction was 30%. These findings were confirmed by magnetic resonance imaging (MRI; Videos 1 and 2), which also demonstrated cardiac fibrosis (Fig. 2). Because of the presence of both a tumour and hypertrophic cardiomyopathy, metaiodobenzylguanidine (MIBG) and octreotide scintigraphy were performed. These showed high fixation of the tumour, consistent with a paraganglioma (Fig. 1B). No other tumour foci were identified. Urine analysis confirmed catecholamine levels five to 10 times normal values and genetic testing demonstrated a characteristic mutation for paraganglioma (SDHC gene).

Unfortunately, because of the location and extensive size of the tumour, only palliative treatment for hydrocephaly with a ventriculoperitoneal shunt, and medical therapy for cardiomyopathy with alpha- and beta-adrenergic blockers, were pursued. Genetic screening was proposed to the patient's family.

The prevalence of paraganglioma is one in 30,000; 30% of patients have a genetic substrate and 10% of tumours secrete catecholamines. The diagnosis is established by MIBG or octreotide scintigraphy; biopsies are not indicated. MRI and computed tomography

Abbreviations: LV, Left ventricular; MIBG, Metaiodobenzylguanidine; MRI, Magnetic resonance imaging.

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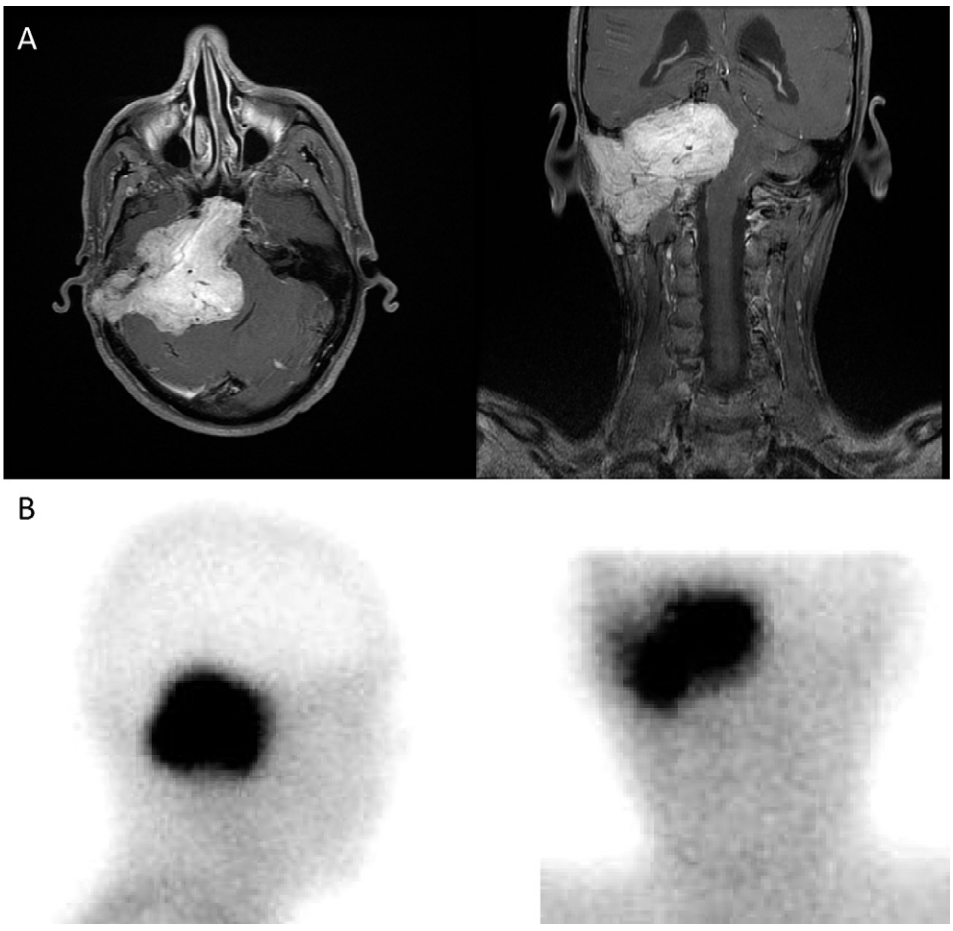

Figure 1. Panel A: cerebral magnetic resonance imaging, T1-weighted, axial and coronal views, showing the tumour with compression of the brain stem and extension to the posterior fossa. Panel B: metaiodobenzylguanidine scintigraphy showing the high fixation of the tumour, consistent with a paraganglioma.

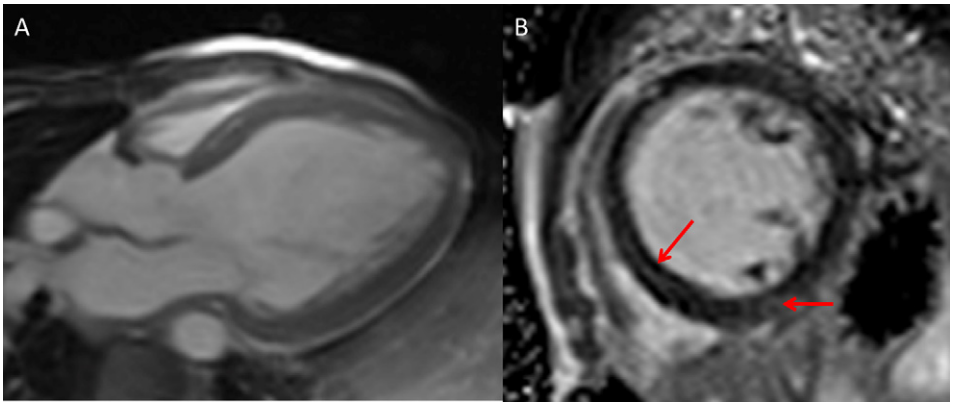

Figure 2. Cardiac magnetic resonance imaging. Panel A: parasternal long-axis view of the left ventricle showing its dilatation and global hypertrophy. Panel B: phase-sensitive infrared single-shot sequence showing linear and nodular intramyocardial enhancement (arrows), consistent with non-ischaemic pattern of myocardial fibrosis.

determine tumour extension and vascularization to guide treatment: embolization, surgery and/or metabolic radiotherapy. Severe hypertension in a young subject, especially if associated with cardiac function impairment and hypertrophy, justifies exhaustive investigations looking for an extracardiovascular aetiology, whereas any patient with a known tumour such as paraganglioma or pheochromocytma should undergo complete cardiac examination before any surgical treatment.

Conflict of interest statement

No conflict of interest to declare.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.acvd.2010.07.004.